THE METASTASIS OF HYPERNEPHROMA IN THE NERVOUS SYSTEM: JACKSONIAN EPILEPSY CAUSED BY SUCH LESION.[1]

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That tumors of the adrenals may exist without causing any symptoms is well known, and that metastasis of such tumors may occur in parts of the body remote from the adrenals, without previous or coincident manifestations of disease in the adrenals, is equally well known. The neurologist has small encounter with hypernephroma metastases. Its occurrence in the central nervous system is uncommon. Occasionally, he has to distinguish bone metastasis of it from brachial, pelvic, and other forms of neuralgia, but even this is of rare occurrence.

We have recently had a case of Jacksonian epilepsy, which was found to be dependent upon a metastasis of hypernephroma. The case presents many interesting features, not the least of which is the extraordinary absence of objective signs to indicate the existence of extensive and advanced disease.

The patient was a Bohemian cigarmaker, aged forty-five years, who maintained that aside from an attack of rheumatism, which he had when he was aged forty-one years, he had not been ill since childhood, until he had an attack of unconsciousness four months before he entered the Neurological Institute in July, 1911. The first attack came on suddenly while he was at work, May 9, 1911. He felt a twitching of the left thumb and forefinger, followed by a sensation of numbness and tension, gradually extending up to the shoulder, and then he lost consciousness. Within an hour he felt quite well again, save that he was weak and discouraged. Five hours later he had a second attack, but without loss of consciousness. A few days after this experience he began to have attacks characterized by paresthesia in the left hand and forearm and twitching of the thumb and index finger, which were not accompanied by any disturbance of consciousness, and which were not followed by any loss of dexterity. He had had from one to five such attacks every day for three months following the original attack. The next severe attack that he had was July 13, 1911, he being at that time in the hospital. He had two severe attacks in one day, each lasting about fifteen minutes, and each followed by a profound sleep. The attacks were typical of Jacksonian epilepsy, the attack beginning as above described and followed by generalized convulsions. A physical examination at this time failed to reveal any signs of organic disease of the brain, nor did the patient complain of any symptoms, save those that have been enumerated. There was no headache, vertigo, disturbance of equilibrium, loss of power in any of the extremities, or disordered function of any of the special senses. He complained only of general weakness, loss of flesh, and of the attacks of twitching or convulsions in the left hand. The attacks were not always the same. At times the sensation in the hand was that of pins and needles, and the twitching might be so slight as to flex only the thumb rhythmically a few times. At other times the sensation in the fingers and hand, wrist, and forearm was of tingling, numbness, tension, and bewilderment, and the motor symptoms were correspondingly greater. The duration of the attacks was from a few seconds to several minutes. He gave up his work because of the frequency of the attacks, and because he felt generally weak. Moreover, the left hand was awkward and weak. He remained in the hospital for one month, during this time he had no major attack, save the one mentioned above.

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[1] Read before the American Neurological Association, June 1, 1912.

He reëntered the hospital September 12, 1911. He had by this time developed the only symptom which, aside from the Jacksonian attacks, he ever developed—namely, mild somnolency. He had likewise developed a physical sign which was striking. The left hand, as he lay in bed, assumed a semi-flexed position, with the thumb extended. The hand had the appearance of being atrophied, though no distinct atrophy could be made out; he could move the fingers freely and readily.

Physical examination showed moderate weakness of the left side of the body, upper and lower extremity, and face. The tendon jerks of the left extremities were livelier than those of the right. There was no Babinski phenomenon and no clonus. The abdominal and epigastric reflexes were elicitable on both sides, those on the right side being somewhat livelier than those on the left. There was slight incoördination of the left hand and a distinct impairment of dexterity of this hand. Tactile sensibility, contact sensibility, and pressure sensibility were unimpaired. He did not interpret quickly and correctly postural sensibility in the left hand, forearm, and arm. On several occasions there was distinct astereognosis of the left hand. This symptom, however, was not constant; it would be found one day and not the following day. Wassermann examination of the blood and cerebrospinal fluid was negative. The fluid obtained by lumbar puncture showed no excess of globulin, one cell, and it reduced by Fehling's solution. No abnormality of the thoracic and abdominal viscera could be made out, save that the heart was dilated, its impulse feeble, its sound puerile. The pupils were circular, equal in size, and reacted promptly to light. The only definite physical sign that existed was an absence of the left plantar reflex.

In the latter part of September an entirely new clinical symptom showed itself in the left hand. This consisted of a rhythmical movement of the thumb and index finger, two or three short adductions of the thumb and index finger, followed by flexor movement only of the index finger and thumb. The first or coarser movements were at the rate of one per second, the slighter at the rate of eight per second. The patient was quite conscious of the movement and experienced considerable discomfort from it. This later manifestation was not accompanied by, or associated with, paresthesia, but since the onset of it the localized convulsions in the hand and forearm became more frequent, sometimes occurring as often as every half-hour. It was also noted that the tendon jerks in the left upper extremity had become much exaggerated since the onset of the rhythmical movement. Although the left hand still had a fair amount of strength and the fingers and hands could be moved, he now complained of a sense of great clumsiness in his hand, and postural sensibility became much impaired. He also complained of severe pain in the back of the hand and wrist; he could not further particularize it. There was no disturbance of contact sensibility or thermal sensibility. The patient had no symptoms of increased intracranial pressure save the somnolency above mentioned. On September 26 it was noted that the rhythmical movement of the thumb and index finger was less in intensity and frequency. At this time the rate was fifty to the minute. On September 25 it was noted that he had three major attacks three hours apart, each of which lasted ninety seconds. The rhythmical movement ceased for one-half hour after each attack. On both of these dates and on September 28 it was recorded in the clinical notes that there was no astereognosis.

Five months after the initial symptom—namely, on October 1, the patient, while lying in bed, had a series of convulsions and died.

To recapitulate: A man, aged forty-five years, who had been in good health developed typical Jacksonian epilepsy, displaying itself by twitching of the left thumb and index finger, paresthesia of the left hand, and forearm and followed on five occasions by loss of consciousness and generalized convulsions. Gradually there developed loss of power and of dexterity in the left upper extremity and to a slight degree in the entire left half of the body. The only definite physical signs were loss of sense of position in the left hand and fingers and absence of the left plantar reflex. There was an inconstant astereognosis of the left hand and a mild increase of the tendon jerks of the left half of the body. A week before his death a slow rhythmical tremor of the left thumb and index finger, consisting of adductor movements about one per second, developed.

When the abdominal cavity was opened a large whitish tumor was found in the right flank. This tumor had invaded the upper pole of the right kidney, surrounded the adrenal and extended into the under surface of the right lobe of the liver. In various other parts of the liver there were a few metastases up to three-fourths of an inch in diameter. In the wall of the duodenum, near the junction with the jejunum, was a small metastasis one-eighth of an inch in diameter. The only other metastasis found was in the right cerebral hemisphere. This metsatasis was one and one-half inches in diameter, situated one-fourth of an inch below the surface midway between the vertex and the base, that is, in the arm area, and more particularly in the hand area.

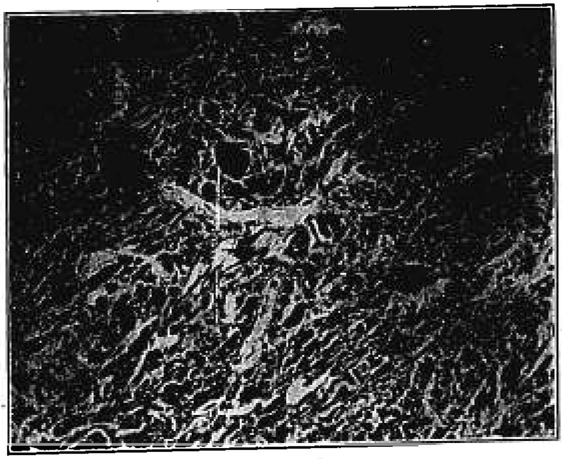

FIG. 1.—Hypernephroma.

Microscopically, the primary tumor mass, as will be seen in the photomicrographs, consisted of cells running in columns arranged about capillaries, the appearance resembling closely that of the cortex of the adrenal gland. The nuclei stain deeply, but the protoplasm stains only lightly with eosin and shows numerous vacuolations. Here and there in the strands are seen some cells much larger in size than the majority, taking a deeper eosin stain and exhibiting granular degeneration or fragmentation of their nuclei (Fig. 1).

In the metastases in the liver the general resemblance to the adrenal cortex is retained, but is interrupted frequently by indecriminate masses of cells with no particular arrangement, and by the presence of a greater number of the larger degenerating cells (Fig. 2).

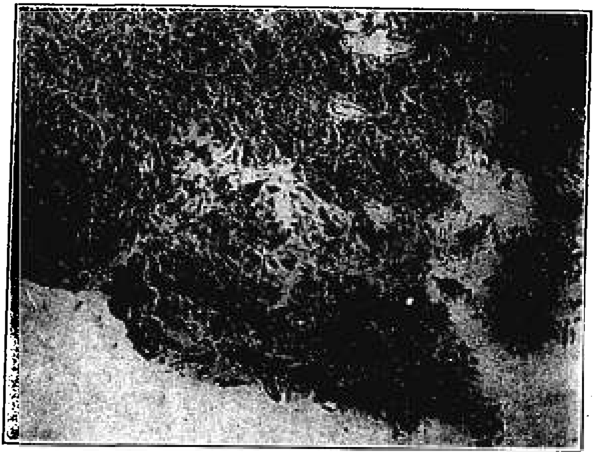

FIG. 2.—Hypernephroma. Metastases in liver.

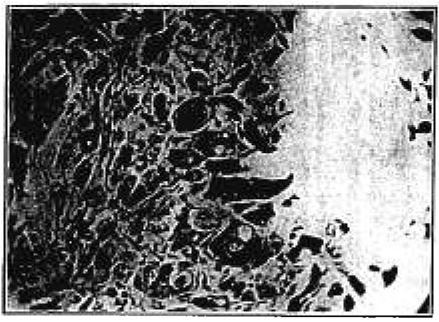

FIG. 3.—Hypernephroma. Metastases in brain.

In the metastasis in the wall of the duodenum the resemblance to adrenal tissue is maintained.

When, however, we come to look at the metastasis in the brain, all suggestion of adrenal tissue is lost. The cells are large in size, polyhedral in shape, stain deeply, and show nuclei in many stages of degeneration. Some nuclei are small, deeply stained, and show no granulation. Others are large, pale in color, vacuolated, granular, and fragmented (Fig. 3).

Many of the cells in this metastasis have taken on a phagocytic function, and can be seen to have ingested red and white blood cells.

The presence of a tumor of this size in the abdomen, without having given rise to more definite symptoms betraying its presence, seems remarkable.

The abdominal wall was very fat, and though several routine examinations were made, no sign or symptom ever prompted more searching examination.

He had at no time any evidence of hemorrhage into the tumor, such as pain or hematuria, and the only symptom he complained of which is sometimes found with hypernephroma was a frequent aversion to food.

The blood pressure, of course, could be expected to give no information, since the tumor was entirely made up of tissue resembling the cortex of the adrenal gland, and this cortex has never been found to affect the blood pressure in any appreciable manner.

The extraordinary absence of the general symptoms of brain tumor is probably to be explained by the fact that the tumor had not caused considerable increase of intracranial pressure. It was this absence of symptoms that decided us not to submit him to exploratory craniotomy, despite the directness of the localizing sign that existed—namely, the Jacksonian convulsions. The rhythmic tremor that developed shortly before his death is of much interest. It has been generally taught that movements of this character conditioned by intracranial disease are indicated by impulses that arise from, or better said, are coördinated in the thalamus.

Without going deeply into the origin, histology, or dissemination of hypernephroma, we may perhaps say a word or two about this form of tumor. It is one of those which cannot be classed rightly with either the carcinomas or with the sarcomas, and will probably in future be put in a class by itself. It may be benign or malignant; that is, it may exist throughout life confined to its original location and cause no discomfort or menace to the patient's life; or it may be the seat of a hemorrhage, due to its friable composition, and so cause intense pain or even loss of life; or, yet again, it may take on a malignant character and strew the body with metastases.

In its simplest form, as a benign tumor, it can be best studied with regard to its origin. Histologically it consists of strands of cuboid cells arranged in long strands about capillary bloodvessels. This arrangement reminds one forcibly of the appearance of the cortex of the adrenal gland, and this similarity is enhanced by the presence in the cells of large quantities of a double light refracting fat which characterizes the cells of the cortical adrenal cells. This constituent may be found in the cells of even the most remote metastasis, though in the case which we report it has seemed to have disappeared in the brain metastasis.

In addition to this histological resemblance to the adrenal gland the location of the tumor is also most significant. Besides its occurrence in the adrenal gland itself, where normally we find small adenomas of minute size, hypernephromas occur most commonly in the upper pole of the kidney and in the right lobe of the liver on its under surface. These parts are in very close proximity in the embryo, and when we find metastasis also occurring in the ovaries and testicles, organs which take their origin in the same close proximity, though later descending to their respective sites, we can hardly doubt the origin of this tumor. This conception of hypernephromas must lead one to regard it as a congenital phenomenon. Later in life, sometimes, we see it take on an activity signifying an entire change of character. It starts to grow rapidly, to invade surrounding tissues and organs, and to disseminate metastases in many different parts of the body. The most common place to find these secondary tumors is in bone, but we here record a metastasis in the brain, in which location many statistics regarding hypernephromas record only one or two instances.

METABOLIC OBSERVATIONS ON AMYOTONIA CONGENITA.

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Amyotonia congenita, or myatonia congenita, is a rare condition affecting children, aged up to three or four years, and exceptionally later, with a general proximal pseudoparalysis, for which no satisfactory etiology or pathology has been advanced. According to a recent clinical review of the subject by Haberman,[1] about 35 cases had been reported up to 1910. Since then a case has occurred in the service of Dr. J. P. Crozer Griffith at the University Hospital, Philadelphia, and was reported by him.[2] Griffith has

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[1] Amer. Jour. Med. Sci., 1910, cxxxix, 383.

[2] Archiv f. Kinderheilkunde, 1910, liv, 211; Trans. Amer. Ped. Soc., 1910, xxii, 184.